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ABSTRACT

This booklet discusses the impact and treatment of the two craniosynostosis syndromes (Apert and Crouzon), which involve the premature fusion of skull sutures, are usually identified at birth, and require years of treatment. It is noted that children with Apert syndrome may have some degree of mental retardation while children with Crouzon syndrome usually have average intellectual potential but often have specific learning problems. Briefly covered are effects of the syndromes on: hands and feet (Apert syndrome children may not have functional fingers); eyes (usually bulging with reduced vision); hearing (significant loss in 10 to 33 percent of cases); mouth (noticeably malformed palates); speech and language development (frequent problems); and breathing (problems are common). Intervention is discussed in sections on the craniofacial team; surgical treatment of the craniosynostosis; surgical treatment of the midface, hands, and feet; and orthodontic treatment. Other services covered include dental hygiene, speech pathology, psychosocial psychiatry, social work, and education. The booklet ends with a discussion of the child's adjustment to his or her peer group. (DB)

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APERT, CROUZON AND OTHER CRANIOSYNOSTOSIS SYNDROMES

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Apert, Crouzon and Other Craniosynostosis Syndromes

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Introduction

The parents of a baby with one of the craniosynostosis syndromes need information and guidance, but even more important, they need reassurance. The problem may seem overwhelming, but there is expert help available to permit their child to grow up and lead a useful and satisfying life. Some of the problems can be completely solved at present, others only partly, but advances are constantly being made.

This booklet discusses the impact and treatment of *Apert and Crouzon* syndromes. Effective treatment takes many years, involves many specialists, and is completed only in late adolescence.

Craniosynostosis

The skull is designed to protect the brain during its period of very rapid expansion before birth and during infancy. To do this, the skull is made up of many bones connected by sutures or areas of bone production (Fig. 1). The force of the growing brain pushes all of the skull bones apart,

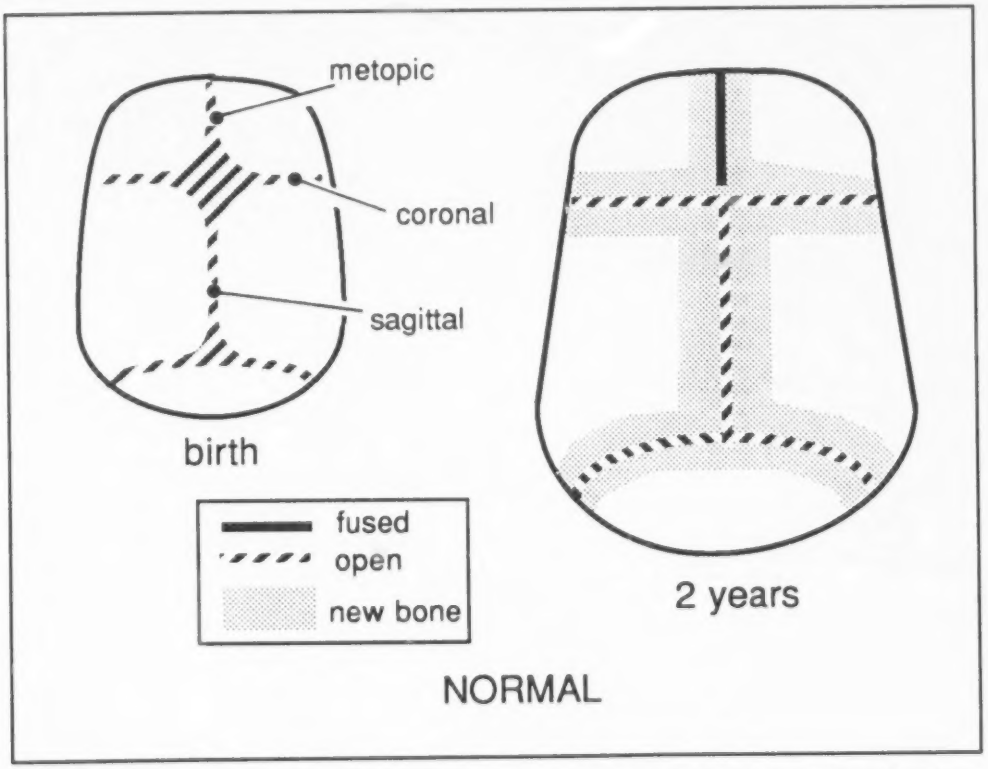

Figure 1: Diagrammatic representation of the top of the skull. This shows the location of the sutures and how the addition of new bone at the sutures permits the skull to enlarge.

The metopic suture (mid-forehead) closes early but the others remain open and growing until the head reaches its full size in adolescence.

causing the sutures to lay down bone to fill in and permitting the smooth, continuous enlargement of the skull. The sutures are oriented to permit growth in different directions. For example, the *coronal* sutures allow the skull to grow in length, while the *sagittal* suture permits increase in width.

Apert and Crouzon syndromes are the most common members of a family of conditions characterized by craniosynostosis, which is the premature fusion of skull sutures. In these conditions one or more sutures of the cranial vault are fused at birth and skull growth is altered.

Once bone fusion has occurred at any point in a suture, bone growth along the entire suture ceases. When one or more sutures fuse in craniosynostosis, the remaining sutures are forced to compensate by extra growth, and distortions in the shape of the skull occur (Fig. 2). Unilateral conditions

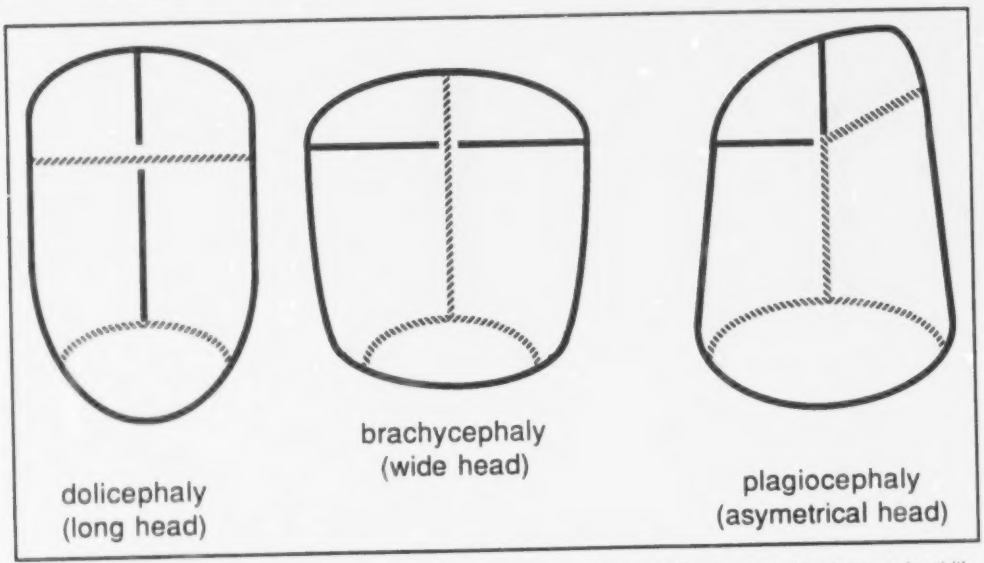

Figure 2: Diagrams of abnormal conditions. When the sagittal suture fuses (left figure), no further increase in width of the skull can occur, and the coronal and lambdoid sutures overgrow to compensate. The head thus becomes long and narrow (dolicephalic). When the coronal sutures fuse (middle figure) no further increase in length occurs, and the sagittal suture overgrows to compensate. The head thus becomes wide (and high from other sutural compensations), a condition termed brachycephaly. When only one of two paired sutures fuse (right figure), overgrowth of the other causes asymmetry, termed plagiocephaly.

(plagiocephaly) result in skull and face asymmetries which are particularly disfiguring. If too many sutures fuse, the skull resists enlargement and intracranial pressure rises. The severity of the problem increases dramatically with the number of sutures affected. For children in whom most or all sutures are fused, the results can be devastating if appropriate treatment is not given early. There may be significant skull anomalies as well as varying degrees of vision and hearing loss and even a reduction in intellectual ability.

In many of these conditions there are further growth problems that develop when the cranial base and midface sutures are partially synostosed. The lower jaw is not directly involved in these conditions.

In most cases, craniosynostosis occurs before birth and is recognizable in the newborn. Why it develops is not known but several different mechanisms may be responsible. In experimental animals, pressure from wedging of the head in the mother's pelvis during late pregnancy has been shown to cause synostosis, as have massive doses of vitamin D. Often the condition is familial, however, and an autosomal dominant transmission is usual. About 50% of children of an affected parent are affected, but sporadic cases also occur frequently. In some patients craniosynostosis develops after birth. It may be related to lack of brain growth, an unusually small head, shunted hydrocephalus, trauma to the skull sutures, metabolic irregularities or progressive synostosis as found occasionally in Crouzon syndrome.

Many of the syndromes in this group involve hand and foot anomalies, which have given rise to many different syndromes being identified on the basis of specific limb anomalies. Crouzon syndrome has no such anomalies while Apert syndrome has severe syndactyly (webbing of the hands and feet).

Today the future for all infants with craniosynostosis is greatly improved. Treatment is available from craniofacial teams made up of various health care personnel. To achieve the best results, patients should be referred to a craniofacial team in their first few months of life.

Apert and Crouzon Syndromes

In addition to premature fusion of sutures in the cranial vault, infants with Apert or Crouzon syndrome have premature fusion of the bones in the cranial base and mid face. The bones forming the orbit are affected and the orbit is usually small and shallow.

Figure 3: Girl with Crouzon Syndrome, showing the characteristic facial features. Severe growth deficiency of the midface from fusion of facial sutures results in a depressed nasal base, although the nose itself is not small; shallow bony orbits which make the eyes appear large and protrusive. The mandible (lower jaw) is normal but appears large by comparison.

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The upper jaw is deficient at birth and thereafter grows at only half the normal rate. Without treatment, the face becomes progressively more flat or concave as the lower jaw grows normally and leaves the upper jaw behind (Fig. 3). Usually the overall height of the face is normal but the vertical proportions are poor because the midface is reduced in height and the lower face has increased height. The nose appears very small, usually because there is too little underlying bone, but also because the face is often very wide. In addition to the face and skull anomalies, these conditions involve other problems as outlined in the following sections.

Intelligence

Children with Apert syndrome may have some degree of mental retardation, although some have normal intelligence. Children with Crouzon syndrome usually have average intellectual potential but often have specific learning problems that require early identification and remedial therapy.

Hands and feet

Children with Apert syndrome have syndactyly or lack of normal separation between the fingers and toes (Fig. 4). The digits may be joined only by skin and soft tissue or by bone as well. As a result, the fingers may not be functional but there is no loss of function in the feet.

Figure 4: Hand of a child with Apert Syndrome where the fingers are fused together, either by skin only or sometimes by bone as well.

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The syndactyly in the hand is most severe among the index, long and ring fingers and less severe between the thumb and index finger and between the ring and little fingers. Often the bones at the tips of the long and ring fingers are fused. In addition, the hand may be turned so that the palm does not face in the normal direction. In the foot, all of the toes are joined by soft tissue, with the 2nd, 3rd and 4th toes being the most severely affected. Often, the first and second toes are also fused.

Eyes

A variety of eye problems are common in patients with Crouzon or Apert syndrome. The eyes usually bulge because the bony orbits are too small to contain them. The lids may not cover the corneas adequately and as a result, the exposed eyes may become too dry and are vulnerable to trauma.

Vision may be reduced in one or both eyes as a result of optical error which may be corrected with glasses or in some cases occlusion treatment or patching. Some patients have strabismus (crossed eyes or wall eyes) which not only presents a cosmetic problem but also may be associated with lazy eye or poor vision. In these cases correction may be achieved with glasses, patching to improve vision and in some cases, surgery to straighten the eyes.

These patients may also have their vision jeopardized by increased intracranial pressure that leads to slow, progressive damage to the optic nerve. The eye examination can sometimes provide the clue for the necessity of early neurosurgical treatment.

Many of the eye problems associated with cranial synostosis can be minimized or treated with the help of an ophthalmologist. In general, the earlier care is begun, the better the final outlook.

Hearing

Hearing loss may be sensorineural (nerve) or conductive. Significant conductive hearing losses occur in over ten percent of patients with Crouzon syndrome and about one-third of those with Apert syndrome because of the fixation of the middle ear structures. Reversible conductive hearing loss is also common. Fluid builds up in the middle ear (i.e., serous otitis media) as a result of skull abnormalities. This impedes the function of the eustachian tube which conducts air from the back of the nose to the ear and normally drains excess fluid. The problem may be corrected surgically by inserting a ventilation tube into the eardrum (tympanic membrane). Although this procedure is considered minor surgery, it must be done with care in patients with craniofacial disorders because the jugular vein may not be covered with bone as is normally the case.

Mouth

Children with Apert or Crouzon syndrome have noticeably malformed palates (Fig. 5A). The midline is very high, since it is attached to an inadequate nasal septum. As the tooth-bearing 'horseshoe' of the maxilla (upper

Figure 5(A): The maxilla (upper jaw) is restricted in width and forward growth. The palate is very high in the midline because the nasal septum is deficient. This becomes worse with growth, since the teeth and surrounding alveolar bone continue to grow downward normally while the midline palate does not. The small maxilla leads to severe crowding of the teeth: in this case three teeth are missing and crowding is still present. The second molars have no space in which to erupt at the back of the dental arch.

jaw) grows downward, the palate becomes increasingly high and narrow. In addition, there are deposits of material on the sides of the palate (mucopolysaccharides) which exaggerate the situation. Although dental development is usually normal, the teeth are extremely crowded in the small maxilla and some erupt into the palate. These features may give the appearance of a cleft in the middle of the palate but in fact there is usually only a very deep groove (Fig. 5B). The hard palate is not cleft unless it is a continuation of a soft palate cleft. Invariably the soft palate is long, hanging down into the pharynx and causing speech and breathing problems.

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Figure 5(B): Often there are masses of material present in the palate which reduces the palatal vault to a narrow slit. Cleft palate or submucous cleft palate is often present, especially in Apert Syndrome.

When the jaws are closed, there is often an anterior open bite (a space between the upper and lower front teeth when the back teeth are clenched). This is due partly to the maxilla being tipped up at the front and partly to the position of the tongue between the front teeth (Fig. 6). The normal tongue cannot occupy its natural position in the vault of the palate because of the reduced size and altered shape of the palate.

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Figure 6(A): There is frequently a large vertical opening between the front teeth when the back teeth are together. This is mainly caused by the small maxilla and narrow palate which create a lack of space for the tongue inside the mouth, thus forcing the tongue to protrude for comfort and to protect the airway.

Figure 6(B): The small maxilla results in a poor relation with the normal mandible. The condition here and in figure 6(A) cause poor chewing patterns, drooling, mouth breathing and faulty speech.

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Speech and language development

The abnormalities of the skull, jaws, teeth and other oral structures frequently lead to speech and language problems. Since breathing through the nose is difficult because of the small nasal airway, small upper jaw and long soft palate, speech resonance is often hyponasal, that is, not enough air passes through the nose during speech. The poor relationship between the teeth and jaws and the shape of the palate interfere with the production of sounds made with the lips and tongue in the anterior portion of the mouth (Fig. 7).

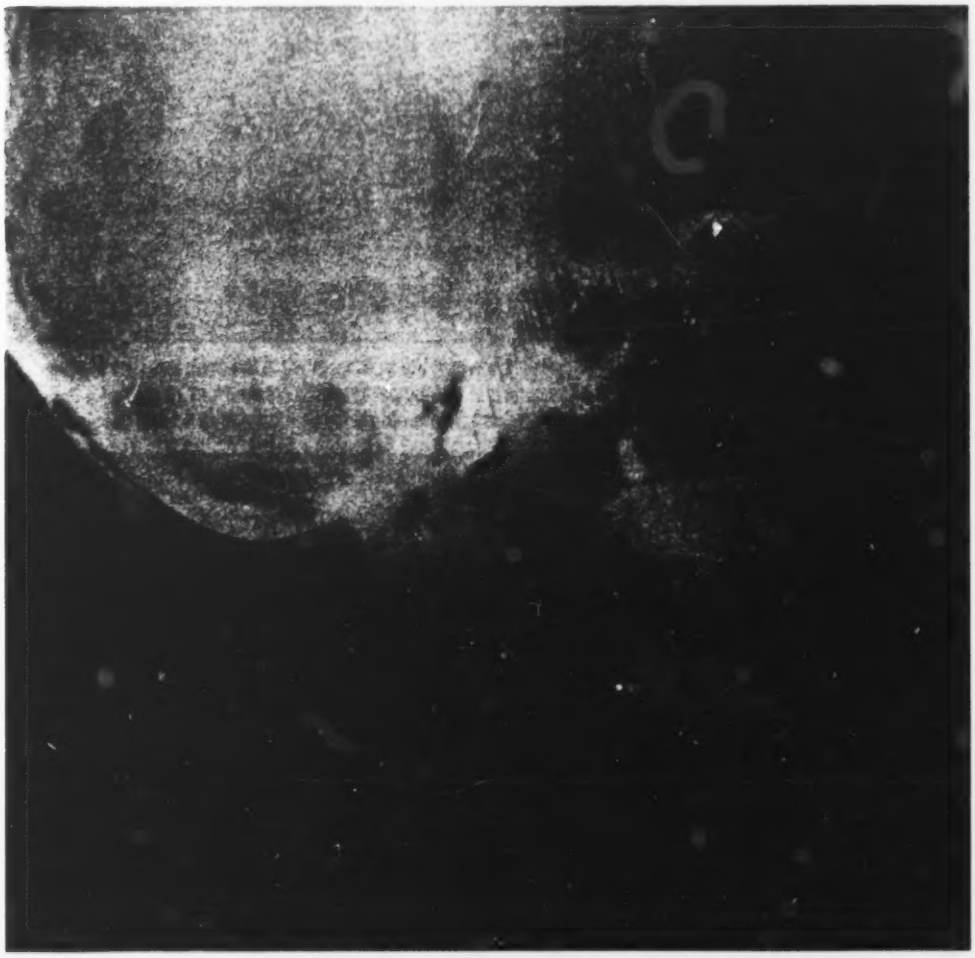

Figure 7: X-ray showing the skeletal dysharmony in Crouzon Syndrome. The long, thick, soft palate partially obstructs the airway and interferes with respiration as well as speech.

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If there are problems with hearing and intelligence, these will contribute to speech and language difficulties. Early parent counselling and speech therapy intervention can help alleviate some of these problems.

Breathing

Breathing problems are common and range from troublesome to life threatening. Newborn infants must breathe through their noses. For many infants with Apert or Crouzon syndrome this becomes difficult because of midfacial retrusion; high arched palate; long soft palate; small nose and anomalies at the base of the skull. Coordination of swallowing and breathing is essential, so for a mouth-breathing infant, modification in feeding techniques or an appliance may be necessary. Severe distress in the newborn period can be relieved by taping an airway in place. Usually any danger of suffocating passes within a few months.

Breathing problems may continue during the first five years of life, especially during sleep. Some children should undergo evaluation in a sleep laboratory. Surgery to relieve continuing troublesome or life-threatening obstructions may include removal of the tonsils and adenoids or moving the midface forward and opening up the airway. In the most severe cases a tracheotomy may be necessary. The decision is made by the craniofacial surgeon in consultation with the otolaryngologist and anaesthetist.

The Craniofacial Team

Since Apert and Crouzon syndromes present such complex and variable problems, treatment is best provided by a craniofacial team, a multidisciplinary group of health professionals usually headed by a craniofacial surgeon, who work together to plan and carry out treatment specifically designed for each patient. A large team might consist of an *anaesthetist, artist, audiologist, facial morphologist, geneticist, neurosurgeon, occupational therapist, ophthalmologist, orthodontist, otolaryngologist, paediatric dentist, paediatrician, psychiatrist, psycho-educational consultant, radiologist, social worker and speech pathologist*..

Since the problems associated with craniofacial malformations affect the child in many ways and also have a major impact on the family, it is essential to treat the whole child in the context of the family and society. The different health professionals work together not only to give each patient the best possible physical treatment, but to encourage positive psychosocial development. They also work with families to help them cope with their own lives while meeting the needs of the affected child.

Treatment

Surgical treatment of craniosynostosis

Treatment begins very early for children with Apert or Crouzon syndrome. Children with craniosynostosis should see the neurosurgeon and craniofacial surgeon within the first few months of life for surgery to open the cranial sutures and allow for normal brain growth.

The application of newer craniofacial surgical principles to the treatment of Apert, Crouzon and other craniosynostosis syndromes has been a major advance in the normalization of both the function and esthetics of these children.

The classical neurosurgical approach of removing the fused suture may achieve a satisfactory cerebral decompression (i.e., pressure relief), but often ends with either rapid or incomplete re-ossification, and usually fails to provide a satisfactory shape to the skull. Ideal treatment therefore demands the cooperative skills of a pediatric neurosurgeon and craniofacial surgeon to simultaneously decompress the brain and reshape the cranial and orbital bones in as normal a form as possible. Figure 8 illustrates some of the surgical procedures used.

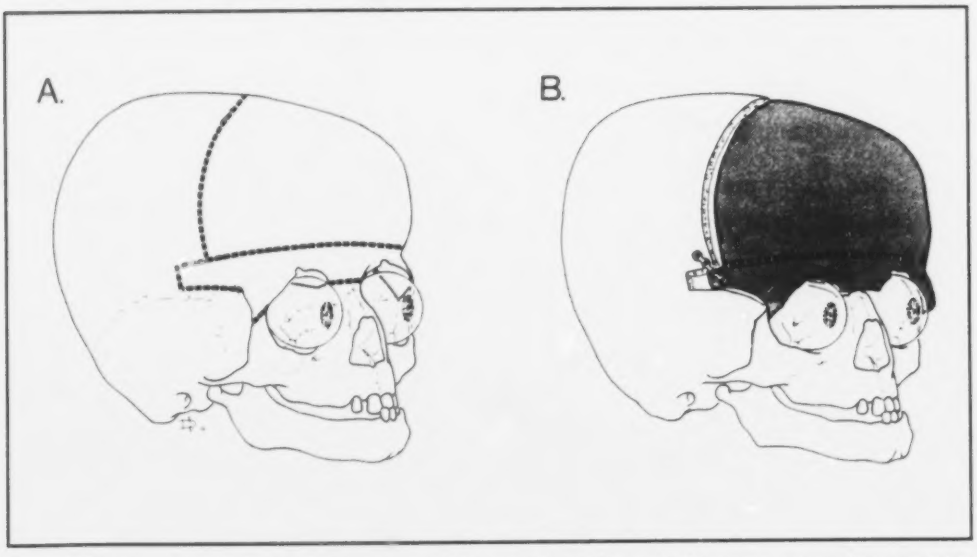

Figure 8(A): Illustration of a skull with hypoplastic midface, shallow orbits and bulging eyes, recessed supra-orbital ridge and slightly bulging forehead (typical findings of Apert and Crouzon Syndromes).

Figure 8(B): Illustration demonstrating supra-orbital ridge osteotomy and advancement stabilized with small bone plates. The forehead has been advanced as well.

Each affected child is unique. Even children with the same basic deformity require individualization of both the type of surgery performed and the timing. Usually, any needed cranial and orbital surgery should be completed within the first year, since the brain almost triples in size during this period. If the rapidly expanding brain cannot find room to grow because of a non-growing cranium, serious problems arise, including increased intracranial pressure with brain atrophy and papilloedema with loss of vision.

The basic initial surgical approach is cranial vault reshaping and supra-orbital bar advancement. Through 'hidden' incisions in the hair-bearing scalp, the top of the skull and forehead are exposed. Next, the appropriate bone of the skull and orbits is removed, while carefully protecting the brain and eyes from potential injury. The bone is reshaped to match the normal appearance as closely as possible. It is often necessary to use bone from other parts of the cranium as free bone grafts to augment deficient areas along the front part of the orbit and cranium, since it is critical to maximize the esthetics in this region.

The child enters the hospital 1 or 2 days before surgery. The operation takes 3-10 hours. The first night after surgery is spent in the Intensive Care Unit and after this the patient may return to the regular hospital ward and be cared for by nurses who are well versed in looking after craniofacial patients. The skin stitches or staples are removed at 7-14 days. In many cases, hospitalization is necessary during this interval. When the child is discharged from hospital, special instructions are given for activity, positioning and signs and symptoms that might indicate problems.

After healing has occurred, the craniofacial team should continue seeing the child at regular intervals to assess progress, growth and development. In ten to twenty percent of infants, it may be necessary to repeat the original operation, either because of premature re-ossification of the suture lines or because the deformity is so severe that a staged approach is needed. In these cases the same hospital course should be expected with each surgical procedure.

Surgical treatment of the midface

The timing and extent of further surgery to the middle portion of the face (upper jaw, orbits, nose and cheekbones) depends on many factors which will be determined by the craniofacial surgeon and the orthodontist. Most specialists prefer to wait until 5-7 years of age. Significant improvement in function and esthetics can be made with surgery before the child enters school where teasing by his peers can cause serious psychological difficulties.

During the operation on the mid face, it is necessary to work in both the nose and the mouth (Fig. 9 and 10). A tracheostomy may be needed on

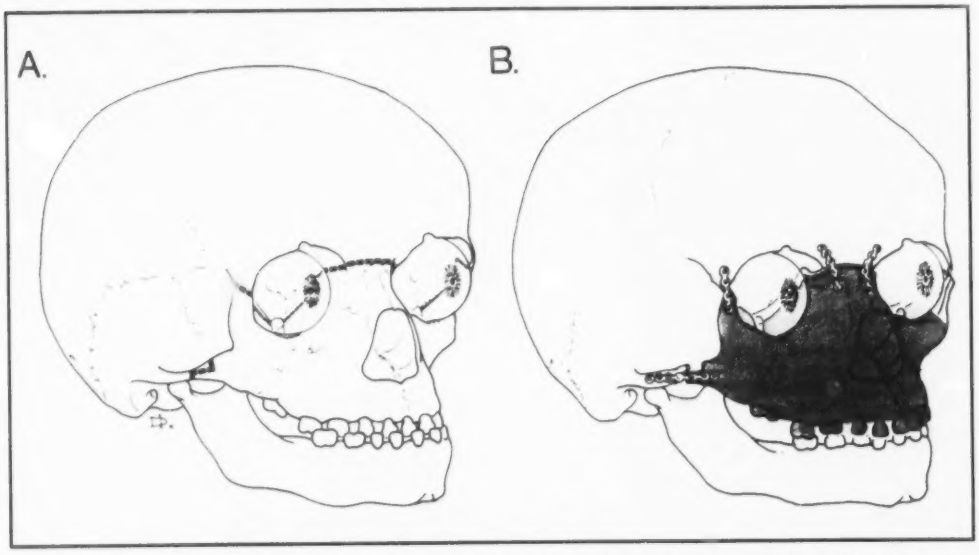

Figure 9(A): Illustration of a six-year-old skull demonstrating midface hypoplasia, shallow orbits and bulging eyes. The supra-orbital ridge and cranial vault are normal. This is often seen in Apert or Crouson Syndrome after the cranial fault and supra-orbital ridge have been corrected in infancy but midface complex remains deficient. Dotted lines indicate osteotomy sites for Le Fort III total midface advancement.

Figure 9(B): Illustration showing Le Fort III total midface advancement and stabilization with metal plates and screws. The eyeballs now sit better in their sockets and the bite has improved.

rare occasions to protect the airway during surgery; this is removed several weeks later and has no permanent effect on the larynx. Repositioning of the upper jaw is done through the previous scalp incisions and in the mouth so that there are no scars on the face. Bone grafts may be needed; these are taken from the skull, rib or hip and do not cause any long-term problems at the site of removal. The repositioned bones are stabilized by wires through the bone or by bone plates and screws. Sometimes the jaws are wired together for 6-8 weeks. The method of stabilization depends on many factors and is determined during the operation.

Figure 9(C): Before early midface surgery.

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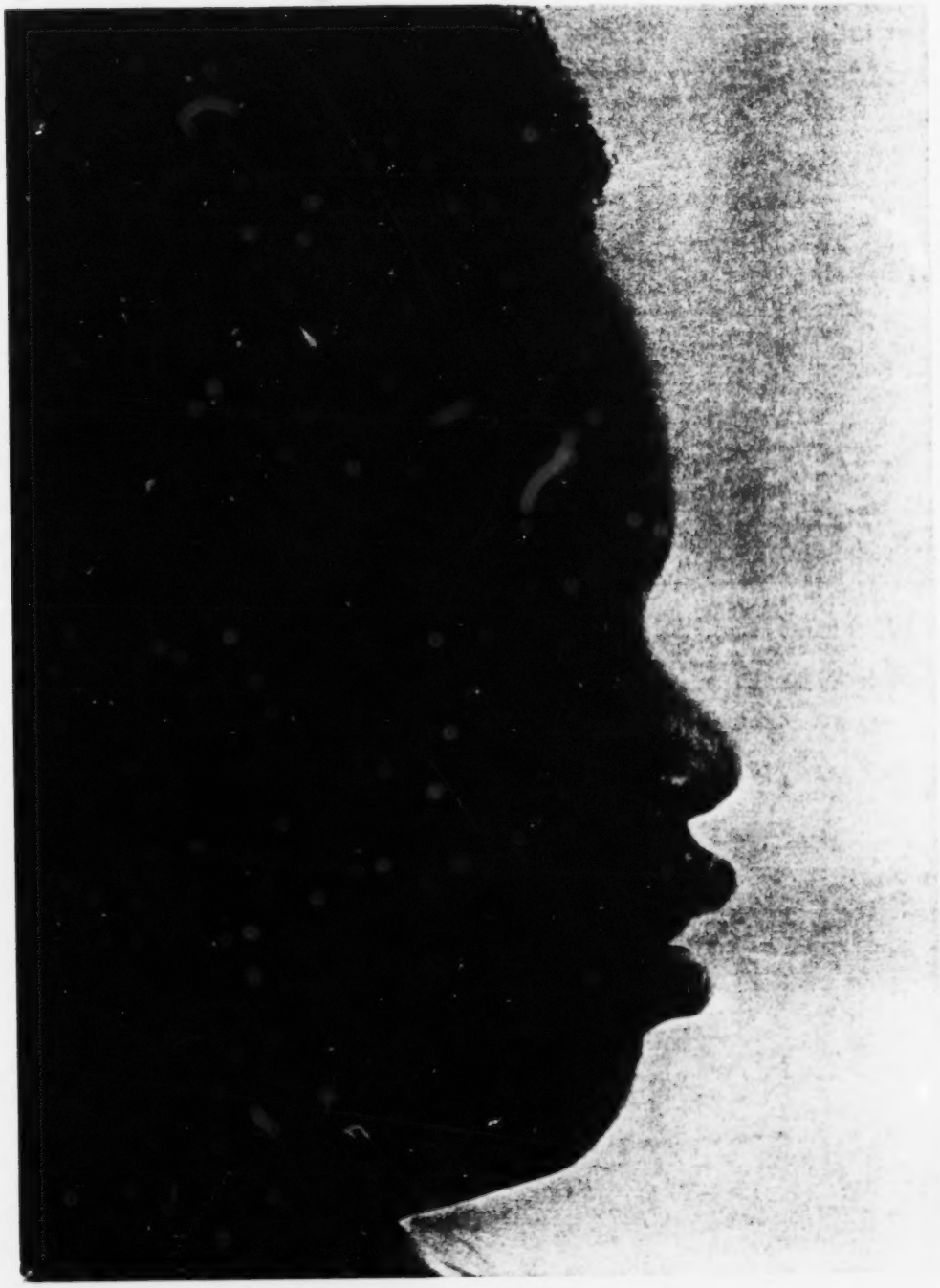

9(D) After: Results of the early midface surgery.

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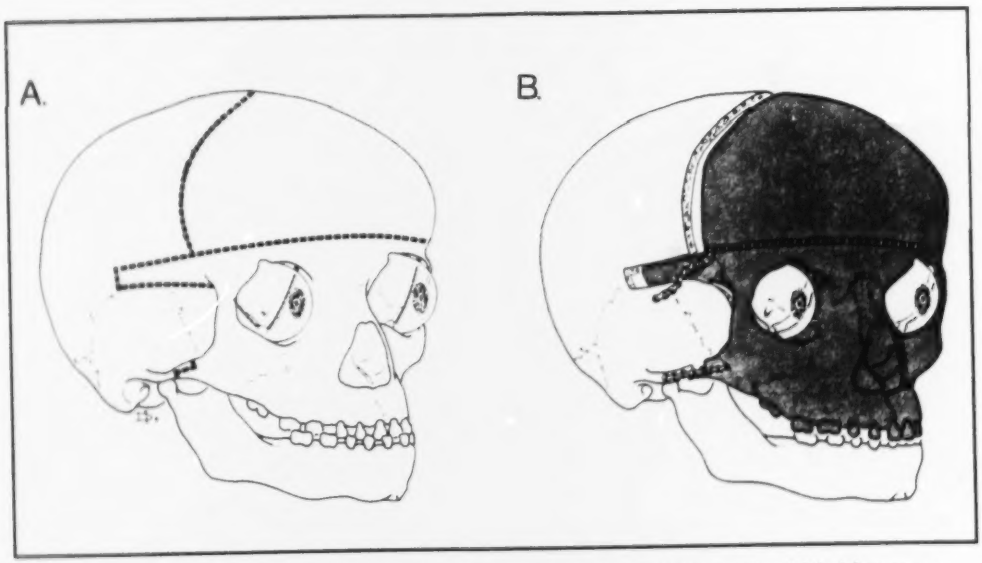

Figure 10(A). The skull of a six-year-old demonstrating midface hypoplasia, shallow orbits with bulging eyes, r esidual flatness to the supra-orbital ridge and residual flatness to the forehead region. Despite previous cranial vault and supra-orbital ridge surgery having been carried out in infancy the supra-orbital ridge and forehead remain flat. The dotted lines indicate the osteotomy sites for the monobloc advancement (total midface and orbital osteotomy) and forehead advancement.

Figure 10(B). Monobloc advancement of midface and orbits completed with improved occlusion and increased orbital depth so that eyes sit in sockets better. The osteotomy is stabilized with small metal plates and screws. The forehead bone has also been advanced forward to give better shape to the cranial vault.

Depending on when the initial surgery is performed, additional operations may be necessary. After the initial surgery the midface does not grow well, whereas the rest of the face does. The good facial proportions achieved during surgery will gradually worsen. As a result, it is necessary to do further facial reconstruction after the bones in the face have finished growing. This gives a permanent correction (Fig. 11)

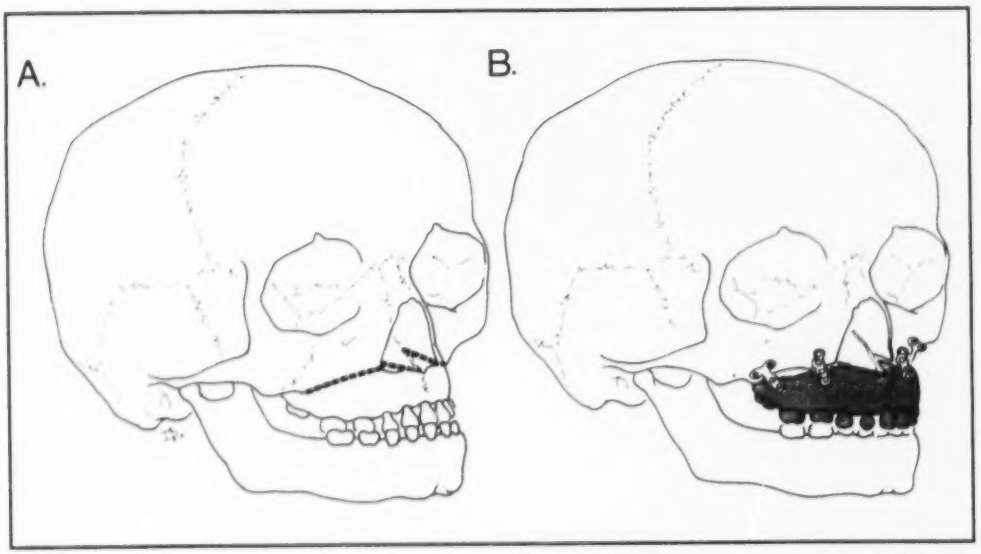

Figure 11(A): The skull of a 16-year-old demonstrating upper jaw hypoplasia resulting in a skeletal dysplasia (jaw discrepancy) and malocclusion (bad bite). Previous surgery carried out in infancy and then early in childhood has corrected problems in the cranial vault, orbits, cheekbones and nose. It is now necessary to "fine-tune" the bite and smile by repositioning the upper jaw through incisions in the mouth.

Figure 11(B): The illustration demonstrates how the upper jaw has been repositioned surgically so that both the bite and smile have improved considerably. The upper jaw osteotomy is stabilized with the use of small bone plates and bone scress to hold the jaw in the new position. It is usually not necessary to wire the jaws together during the recovery period.

Surgical treatment of hands and feet

Treatment of syndactyly in the hands of patients with Apert syndrome involves separating the digits and reconstructing the adjacent surfaces with local skin flaps and skin grafts. Ideally the adjacent webs of any single digit should not be separated at the same operation and therefore two operations are usually done about six months apart. The 2nd and 4th webs can be separated in the first operation and the 1st and 3rd in the second. It is possible to separate all four digits at once, however. The adjacent surfaces are usually repaired using split thickness skin grafts from the buttocks.

The joints of the fingers are not usually functional, except for the meta-carpophalangeal joints and the distal interphalangeal joint of the little finger. The thumb is very short and abnormally positioned. When growth is almost complete, the direction of the thumb can be corrected.

In the feet, surgery is usually simpler and does not involve bone. However, with growth, the heads of the first and second metatarsals may be distorted. As a result, it may be necessary to do an osteotomy to release them and achieve a more normal position for walking.

The timing of surgery is important. The 2nd and 4th webs can be released at about one year of age and the 3rd and 4th webs at about age

two. The position of the thumb can be corrected at age eight and if the metatarsals in the feet need correction, this can also be done at about age eight. Each surgical procedure involves approximately 10 days in hospital as well as a considerable time for rehabilitation after discharge.

We feel it is important to achieve ten digits in both the upper and lower extremities to make them as functional and aesthetically acceptable as possible. We try to avoid amputation of digits even when they are very malformed. By coordinating surgical procedures, we attempt to achieve limbs that are functional for virtually all activities (Fig. 12.)

Figure 12(A): Fused fingers of a child with Apert Syndrome before surgery.

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Figure 12(B): Results of surgery to separate the fused fingers.

Orthodontic treatment

Poor placement and alignment of the teeth must be corrected in conjunction with the craniofacial surgery. The maxilla (upper jaw) is advanced and tipped down to establish a reasonable relationship between the upper and lower jaws. This may be done in early childhood, although surgery at that stage is not a permanent correction since there will be little or no forward growth of the midface after surgery. As the rest of the face continues to grow, the benefit of the original surgery is reduced and a second operation is almost always necessary during adolescence.

Early orthodontic care involves managing the severe crowding of the teeth. The appropriate primary (baby) and secondary (permanent) teeth are extracted to allow for some natural improvement. Braces may be used in children from 7-9 years of age to widen the maxilla or align the front teeth.

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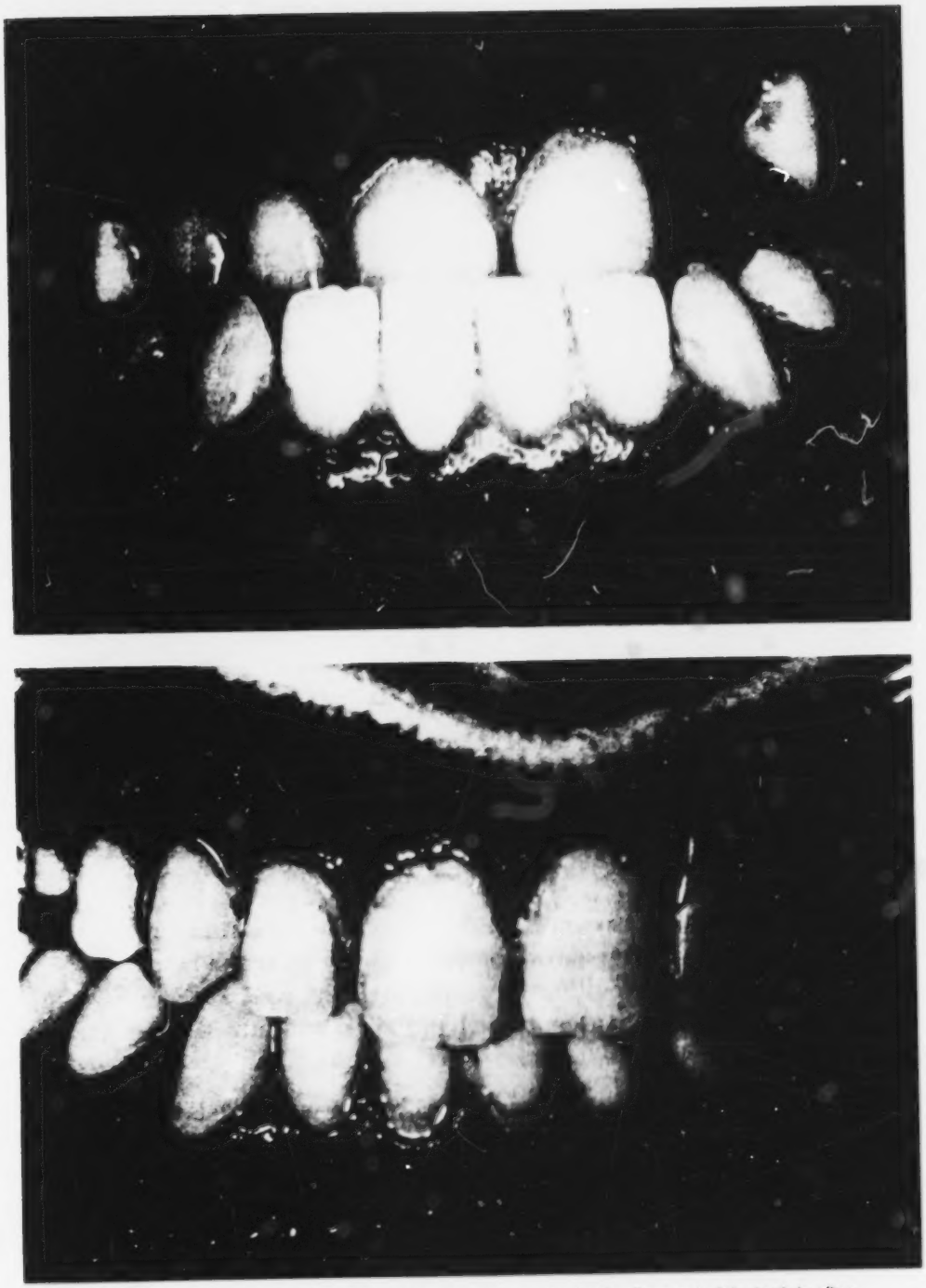

Figure 13(A) & (B): Prior to the final jaw surgery in the teenager, orthodontic alignment of the teeth is often accomplished. Surgical advancement of the maxilla then achieves the result shown (B). A prosthetic (partial denture) replaces the missing teeth on the left side.

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Extensive orthodontic work begins when all of the permanent teeth have erupted (about age 12) and before and after the final jaw surgery (Fig.13). The orthodontist and the craniofacial surgeon plan the details of the final surgery and decide on the appropriate time for it.

Dental hygiene

Throughout the period from infancy to the final correction of the jaw malformations, patients with craniosynostosis must take precautions to avoid dental cavities. Their crowded teeth and the often prolonged wearing of braces make them more prone to cavity formation than other children.

They are encouraged to brush three times a day and use dental floss once a day. In addition, it is recommended that they use a topical fluoride gel once a month if they are having many cavities or are undergoing orthodontic therapy. *Systemic fluorides*, as in drinking water or available in tablets in non-fluoridated areas, offer the best protection for developing teeth and jaws. During orthodontic therapy, an oral irrigator such as a WaterPik is helpful in removing gross food particles but this should not be seen as a substitute for brushing.

Speech and language development

The speech pathologist on the craniofacial team identifies the factors contributing to difficulties in speech and language development and recommends treatment to overcome them. This might involve surgical alteration of the relation of the jaws, orthodontic alignment of the teeth and speech therapy.

Hearing problems associated with craniofacial malformations may delay speech and language development. If children cannot hear sounds accurately, they may have difficulty producing them well. Early evaluation by an audiologist helps to identify such problems and provides information needed by the otolaryngologist and speech pathologist.

Sometimes, mental retardation further contributes to delayed language development. Some children are slow to understand language and put sounds together into words. For example, they may be able to make the "b" sound in "ball" but omit it in "tub." Difficulties in co-ordinating the tongue, lips and teeth also result in poor speech clarity.

The relationship between oral structures and speech is not always predictable. Some children compensate for their *orofacial* anomalies while others do not. Regardless of the child's efforts, however, an underdeveloped midface involves structural problems that may cause distortion of sibilants (s, z, sh, zh), protrusion of the tongue for tongue-tip sounds (t, d, l, n, s, z, sh, ch, zh, j) and a distortion of the 'r' and 'l' sounds. If the jaws do

not close properly, speech sounds requiring lip closure (p, b, m) may be difficult as may 'f' and 'v' sounds. Missing, crowded or rotated teeth may also affect speech.

When air is restricted or prevented from entering the nasal passages, the person's speech sounds as if he has a cold. This is a common problem in craniosynostosis syndromes and is often attributed to the abnormally long soft palate and reduced pharyngeal depth.

Various tests are used to assess the child's speech and the function of the soft palate. In addition, audio- and videotape recordings are made to document the child's speech. These procedures are helpful in indicating whether speech therapy is needed and also in demonstrating the impact of facial surgery on speech. After facial advancement, many children have fewer *articulation* errors and better *resonance* balance.

If speech therapy is indicated, it may be started very early and continued through the child's school years. It must always be based on evidence that the child has the potential to develop better speech and language skills given the existing structural and functional status.

Psychosocial Considerations

The birth of a child with craniofacial malformations represents a major crisis even for the most well adjusted parents. The parents, the infant's siblings, the grandparents and other close relatives may experience shock and grief. Many factors contribute to these feelings.

Some parents feel guilty or angry because they have passed on an inherited problem. Others anticipate pressure from family or society. Many fear the financial implications of the baby's problems. As a result of these anxieties, the marital relationship may be strained and other family members may have difficulty adjusting.

Under these circumstances, families benefit from specialized psychosocial guidance and support, especially during the critical period after birth and diagnosis. They need comprehensive explanations and often welcome preparation for planned corrective surgery and its implications. Usually the parents want to discuss the child's long-term needs. If there are concerns with the child's early development, a psycho-educational consultant can provide the parents with information about early stimulation programmes and activities that can be undertaken at home. Each child is assessed individually in the family context. The psychiatrist, social worker and psycho-educational consultant on the team play a major role in the child's care for several years and provide counselling and advice to the family as well.

The psychiatrist

The psychiatrist's major concerns are the child's future self esteem and social competence. Parents are reassured that the severity of the craniofacial malformation is much less important than the strength of the family in determining how the child will ultimately adjust and succeed in life.

The psychiatrist monitors the patient's self esteem as treatment proceeds through childhood and adolescence, and encourages the development of particular skills and talents. In addition, the psychiatrist helps to build ego strengths and provides positive role models from other patients and famous adults, and ensures that residual physical malformations do not become the focus for other underlying conflicts, such as sexual or familial problems. Individual therapy can be provided as indicated.

Specific psychiatric concerns include adjustment to body image and general self concept as well as ability to relate to family, peers and strangers. Motivation for and expectations of surgery are explored carefully. In the early years, play therapy is often used to help the child integrate the experiences of hospitalization and surgery. In addition, the psychiatrist works with the parents to build on the child's strengths and special abilities.

The social worker

The social worker discusses the child's appearance and diagnosis, and eliminates misconceptions the family may have about facial malformations. In addition, the social worker helps the family with the shock, guilt, anger or depression they may initially feel.

Social workers can act as a liaison between the family and the health care professionals involved in the child's care. They provide specific assistance with surgical planning as it affects the whole family and offer counselling on an individual basis, using a family approach or group sessions, depending on the family's particular needs.

As time goes on, the social workers help parents answer their child's questions about his/her appearance by giving simple, direct, concrete explanations suited to the child's level of maturity. They advise the child on how to handle stares and questions from others on the street or in the playground. They also prepare the child and family for surgery and ensure that they have realistic expectations.

The psycho-educational consultant

Some children with craniofacial malformations have learning problems. The psycho-educational consultant advises parents on early childhood stimulation techniques to encourage optimum development. As the child grows, the consultant assesses the child's learning ability, readiness skills and social maturity and advises the parents on the type of nursery school and any additional stimulation the child may need. If learning difficulties arise, the consultant can recommend specific remedial programmes. Identifying potential problems is of the utmost importance throughout the child's early schooling. Later, the consultant can play a major role in helping individual patients to make suitable vocational decisions.

Adjustments

For young children, 'fitting in' becomes more important than pleasing parents. In the primary grades, children who are different from their peers often have considerable difficulty. They may be teased and called names. Some children learn to cope by educating their classmates. Others rely on their personality strengths such as a quick wit or ability to achieve or by demonstrating specific talents. Self esteem is determined primarily by the ability to feel genuinely positive about one's self or some aspect of one's life, whether it be academic success, sports, music, art or a particular hobby.

During the early school years, many parents are particularly concerned that their child be educated in an environment without pity, overprotection or underestimation of potential. Since teachers are largely unfamiliar with craniofacial problems, it is helpful if parents explain their child's condition. There is a difficult balance to be found between protecting the child from the cruelty of teasing, stares and questions and letting him cope with the world as it is and build ego strengths.

As the child reaches puberty, appealing to the opposite sex becomes important. Virtually all teenagers go through periods of doubt and insecurity. It is not surprising, therefore, that teenagers with a facial defect experience a sharp decline in self confidence. They become more aware of the impact the facial problems may have on their lives, and also of the limitations of treatment. It is a very difficult period but the vast majority manage to cope effectively. Some appear indifferent to the opposite sex and focus on academic or athletic achievements. Life usually becomes much easier when their peers mature and learn to see the person behind the facial appearance.

Conclusion

Apert, Crouzon and the other syndromes with craniosynostosis are complex anomalies with major impact on the lives of those affected. The treatment available at present is imperfect, but it is extraordinary compared to what was available twenty years ago. Hopefully, future advances will be equally dramatic. For the present, however, a large group of health professionals is required to provide treatment that enables affected individuals to live full, satisfying lives.

Notes

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